

Case Report

A Rare Cause of Right Upper Quadrant Pain in a 17-Year-Old Female

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A 17-year-old Hispanic female presented to our hospital with complaints of right upper quadrant abdominal pain, vomiting, and fever. Physical exam was positive for hepatomegaly. Abdominal computed tomography showed multiple hypoechoic liver masses. Liver biopsy was done, which was diagnostic for hepatic epithelioid hemangioendothelioma.

1. Introduction

Hepatic epithelioid hemangioendothelioma (HEH) is an extremely rare malignancy originating from vascular endothelial cells [1]. HEH was first reported by Ishak et al. in 1984, who reported 32 cases of HEH [2]. Unfortunately, HEH lacks specific clinical signs and laboratory markers. The definitive diagnosis is based on the liver biopsy.

Below, we present a case of a 17-year-old female who presented to the hospital with complaints of right upper quadrant abdominal pain. Abdominal imaging revealed multiple liver masses, and biopsy was diagnostic for HEH. We will briefly discuss the available literature on HEH.

2. Case Presentation

A 17-year-old Hispanic female presented to the emergency department with complaints of the right upper quadrant abdominal pain. The pain had worsened over the past three days. Initially the pain was intermittent and later progressed to be constant. Review of symptoms was positive for vomiting and fever. No change in bowel habits or urinary pattern was reported.

On physical examination, the patient had right lower and upper quadrant tenderness with hepatomegaly two centimeters below the right costal margin. Laboratory tests including complete blood count, comprehensive metabolic panel, lipid panel, and pancreatic enzymes were within normal limits.

Abdominal ultrasound was done, which showed multiple hypoechoic masses scattered throughout the liver, measuring up to 2.0 cm in size. Computed tomography (CT) scan showed multiple low attenuation areas in the liver (Figure 1). Liver biopsy was performed, which was consistent with the diagnosis of HEH staining positive for CD31, a known endothelial marker (Figure 2). Skeletal survey and chest CT did not show evidence of metastatic disease.

Given the multifocal hepatic disease in this patient, she was not deemed to be a candidate for liver resection. The patient was referred for liver transplantation and is currently on the waiting list.

3. Discussion

HEH is a very rare liver tumor originating from vascular endothelial cells [1]. The disease pathogenesis is poorly understood. However, an overexpressed vascular endothelial growth factor (VEGF) may be a culprit in HEH [3].

Mean age at presentation is 41.7 years according to a review of 434 cases [4]. Right upper quadrant abdominal pain, weight loss, and liver enlargement are the most common clinical signs encountered in patients with HEH [4]. Lungs, peritoneal cavity, abdominal lymph nodes, and bones were the most commonly reported metastatic sites for HEH [4]. On extremely rare occasions, the tumor may metastasize to the neck [5] and may be clinically presented in adults as

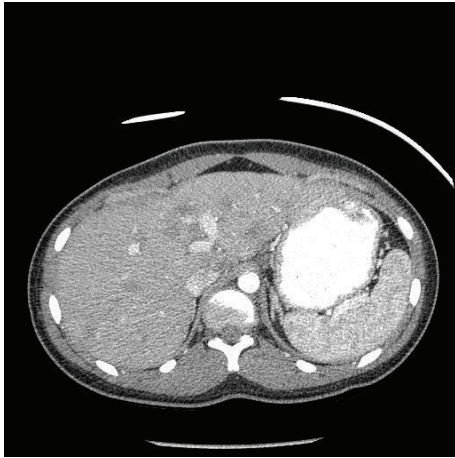


FIGURE 1: Abdominal CT showing multiple poorly defined low attenuation lesions.

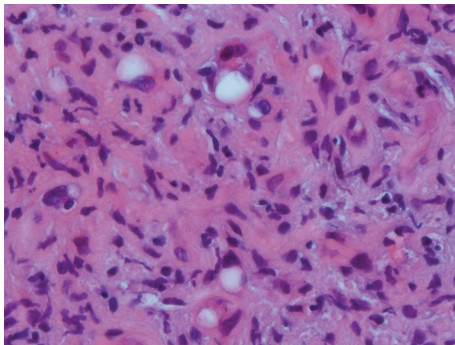


FIGURE 2: Liver biopsy showing scattered epithelioid cells. The tumor cells were stained positive for CD 31.

Kasabach-Merritt syndrome [6], which is a vascular tumor-related thrombocytopenia and bleeding.

Unfortunately, no laboratory biomarker is available for screening. However, cancer biomarker CA19-9, if abnormal, may indicate a poor prognosis in patients with HEH [7]. Abdominal imaging modalities such as CT, Magnetic Resonance Imaging (MRI), and positron emission tomography (PET) may be used in the workup of patients with suspected HEH [8, 9]. Nevertheless, the definitive diagnosis of HEH requires liver biopsy [1]. Histologically the tumor cells are positive for vascular markers such as CD 31, CD 34, and coagulation factor VIII [1].

HEH has a highly variable prognosis ranging from a benign course to rapidly progressive with a poor outcome. Treatment is based on the radiological features of HEH: in a limited disease (less than three liver segments involved) tumor resection can be achieved, whereas liver transplantation is recommended for patient with multifocal liver disease [1]. Transcatheter arterial embolization of the tumor may be considered in selected cases such as in patients awaiting liver transplantation with multifocal liver involvement [4].

Several case studies reported that chemotherapy with radiation therapy may be useful in patient with HEH [4]. Two

recent case studies reported that thalidomide used in patients with HEH led to a clinically and radiologically stable disease after 7 and 9 years of followup, respectively, [10, 11]. In the most recent case report on the treatment of HEH, Sangro et al. reported beneficial effects of VEGF inhibitor drug sorafenib [12]. These reports further support the notion that abnormal and enhanced angiogenesis is likely playing a leading role in the disease pathogenesis.

In conclusion, HEH is a very uncommon disease with nonspecific clinical features. Imaging studies are of some utility, and hepatic biopsy is essential to establish a diagnosis. Given its extremely rare incidence, it is almost impossible to run randomized controlled trials, and the clinicians guide the management based on the published case series.

Conflict of Interests

The authors declare no conflict of interests.

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